



TITLE:

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1 Prenatal diagnosis of anterior sacral meningocele

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15 Keywords; Currarino syndrome, cloacal anomaly, enteric duplication cyst,

16 hydrometrocolpos, MRI

17

1 Abstract

2  
3 Prenatal diagnosis of anterior sacral meningocele (ASM) is an extremely rare  
4 condition and there has been only one English case report posted on the website.

5 A 36-year-old primigravida was referred for huge fetal pelvic cyst noted in routine  
6 ultrasonography (USG) at 19<sup>+4</sup> weeks' gestation. Neither fetal USG nor magnetic  
7 resonance imaging (MRI) at 20<sup>+5</sup> weeks' gestation could detect communication between  
8 the cyst and the spinal cord. Since extension of the pear-shaped cyst through the  
9 pelvic diaphragm down to the perineum was reminiscent of dilated vagina and uterine  
10 cervix, tentative diagnosis of hydrometrocolpos secondary to imperforate hymen had  
11 been considered.

12 In follow-up MRI at 33<sup>+5</sup> weeks' gestation, narrow stalk connecting the pelvic cyst  
13 and the spinal canal through the anterior sacral foramen was clearly delineated, allowing  
14 us to reach the prenatal diagnosis of ASM.

## Case report

A 36-year-old nulligravida became pregnant after in vitro fertilization and embryo transfer. She was referred for huge fetal pelvic cyst noted in routine ultrasonography (USG) at 19<sup>+4</sup> weeks' gestation.

Fetal USG at 20<sup>+5</sup> weeks' gestation showed pear-shaped pelvic cyst and bilateral hydronephrosis with normal amniotic volume (Figure 1A and 1B). Appearance of the external genitalia exhibited normal female phenotype. The pelvic cyst was located posterior to the bladder and extended down to the perineum. The size of the cyst was unchanged during the fetal urination, indicating that the cyst was not communicated with the bladder. Extrinsic ureteral compression by the cyst was considered to be a primary cause of bilateral hydronephrosis. Fetal magnetic resonance image (MRI) taken on the same day revealed that the pelvic cyst with multiloculated nature was located contiguous to the normal-appearing sacrum (Figure 1C). Differential diagnosis included cloacal anomaly, enteric duplication cyst, hydrometrocolpos secondary to imperforate hymen, and anterior sacral meningocele (ASM). Cloacal anomaly seemed unlikely due to lack of communication between the cyst and the bladder<sup>1</sup>. Enteric duplication cyst was implausible because the cyst had no peristaltic movement<sup>2</sup>. Communication of the cyst with spinal canal through sacral bony defect, which is characteristic of ASM, was not detected. Extension of pear-shaped cyst through the pelvic diaphragm down to the perineum was reminiscent of dilated vagina and uterine cervix<sup>3</sup>. Although multiloculated nature of the cyst could not be fully explained, tentative diagnosis of hydrometrocolpos secondary to imperforate hymen was considered.

Diameter of the renal pelvis was relatively constant and amniotic volume was preserved throughout the pregnancy, although the size of the pelvic cyst was gradually increased. The second MRI was taken at 33<sup>+5</sup> weeks' gestation to reevaluate the diagnosis. In the T1-weighted image, the bowel containing hyperintense meconium signal was visualized separately from the cyst (Figure 2A). Narrow stalk connecting the pelvic cyst and the spinal canal through the anterior sacral foramen was clearly delineated in the T2-weighted image (Figure 2B and 2C). Accordingly, we reached the prenatal diagnosis of ASM. Additional meticulous USG search based on the MRI

1 finding, however, failed to visualize the connecting stalk.

2 A female baby weighing 2530 g was vaginally born at 38<sup>+2</sup> weeks' gestation with  
3 normal Apgar score. Her left buttock was bulging with soft cystic mass palpable  
4 beneath the skin. No abnormal neurological finding was noted. Postnatal MRI at 5  
5 days old (Figure 3A) confirmed the communication between the pelvic cyst and the  
6 subarachnoid space through left anterior sacral foramina (S3/4 and S4/5). Multilocular  
7 cyst in the buttock region seemed distinct from the pelvic cyst. Helical computed  
8 tomography at 51 days old (Figure 3B) revealed the defect in posterior aspect of the  
9 vertebral bones below L3. The neonate thrived and remained asymptomatic except for  
10 mild constipation. The operation was performed with posterior approach at 52 days  
11 old. Since the posterior approach failed to identify the pedicle of the pelvic cyst, the  
12 second operation was planned at 87 days old. Both the pelvic cyst and the buttock cyst  
13 were successfully resected with abdominal and perineal approaches, respectively.  
14 Histologically, the wall of the pelvic cyst was lined by ependymal cells and contained  
15 fibrous stroma with neuronal tissue, indicating its meningeal origin. On the other hand,  
16 the buttock cyst was composed of skin, fat, muscle, and mature neural tissue and was  
17 diagnosed as mature cystic teratoma.

## 1 Discussion

2  
3 ASM is herniation of meningeal sac into the presacral retroperitoneal space through a  
4 congenital defect in the sacrum or through the widened anterior sacral foramina <sup>4</sup>.

5 Prenatal diagnosis of ASM is an extremely rare condition and to our knowledge, there  
6 has been only one English case report posted on the website <sup>5</sup>.

7 Currarino syndrome is a unique complex of congenital caudal anomalies. These  
8 include anorectal malformation, sacral bony abnormality, and presacral mass  
9 (meningocele, teratoma, enteric duplication cyst, or any combination of these) <sup>6</sup>.

10 Currarino et al. proposed incomplete separation of the neuroectoderm (future spinal  
11 cord) from the endoderm (future intestinal tract) as its possible embryonic etiology <sup>7</sup>.

12 Half of the cases of Currarino syndrome are inherited <sup>7</sup> and one or two features of the  
13 triad are commonly absent in the same family members, suggesting that this syndrome  
14 should be considered as a spectrum <sup>8</sup>. In this respect, the present case is considered to  
15 fall into the category of incomplete Currarino syndrome.

16 The age and the symptom at first presentation range widely in the patients with  
17 Currarino syndrome <sup>9</sup>. Associated anorectal malformation can cause bowel  
18 obstruction, leading to the early diagnosis in infancy <sup>10</sup>. In the absence of anorectal  
19 malformation, however, most of the patients remain asymptomatic until early adult life.  
20 These patients usually present with symptoms resulting from compression of  
21 surrounding tissue by enlarging presacral mass <sup>11</sup> or acute meningitis <sup>12</sup>. Occasionally,  
22 the incidental discovery of a pelvic mass is the first presentation <sup>4</sup>. Thus, indication for  
23 surgical resection of asymptomatic ASM still remains to be determined.

24 When fetal presacral cyst is encountered, a variety of congenital anomalies including  
25 cloacal anomaly, enteric duplication cyst, hydrometrocolpos secondary to imperforate  
26 hymen, and ASM should be considered. Firstly, exclusion of cloacal anomaly, which  
27 is defined as a confluence of the rectum, vagina, and urethra into a single common  
28 channel, is of great importance because this anomaly usually requires intensive  
29 postnatal care with the worst outcome. For this purpose, one should evaluate possible  
30 communication between the cyst and the surrounding organs. USG is a suitable  
31 measure to see the communication with the bladder, because process of emptying the  
32 bladder into the cyst can be easily visualized <sup>1</sup>. In contrast, USG detection of

1 communication with the bowel may be difficult. T1-weighted MRI can highlight the  
2 meconium and visualization of meconium-containing bowel as a separate structure from  
3 the cyst contributes to exclusion of cloacal anomaly<sup>13, 14</sup>. Secondly, since aspiration of  
4 meningeal cyst could induce chemical meningitis resulting in serious neurological  
5 sequelae<sup>15</sup>, potential communication between the cyst and the spinal canal should be  
6 meticulously evaluated. When the communicating stalk is too narrow for USG  
7 detection as in the present case, MRI may be an essential tool for the prenatal diagnosis  
8 of ASM.

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Figure legend

Figure 1. Coronal (A) and horizontal (B) fetal ultrosonogram and sagittal T2-weighed fetal magnetic resonance image (C) at 19 weeks' gestation.

A. Pear-shaped pelvic cyst (asterisk) is extended down to the perineum.

B. Pelvic cyst (asterisk) is located posterior to the bladder (arrowhead). Extrinsic ureteral compression by the pelvic cyst causes left (L) and right (R) hydronephrosis.

C. Pelvic cyst with multiloculated nature (arrow) is contiguous to the sacrum.

Figure 2. Fetal magnetic resonances images (MRI) at 33 weeks' gestation.

A. Coronal T1-weighted MRI. Bowel containing hyperintense meconium signal (black asterisk) is delineated as a separate structure from pelvic cyst (white asterisk).

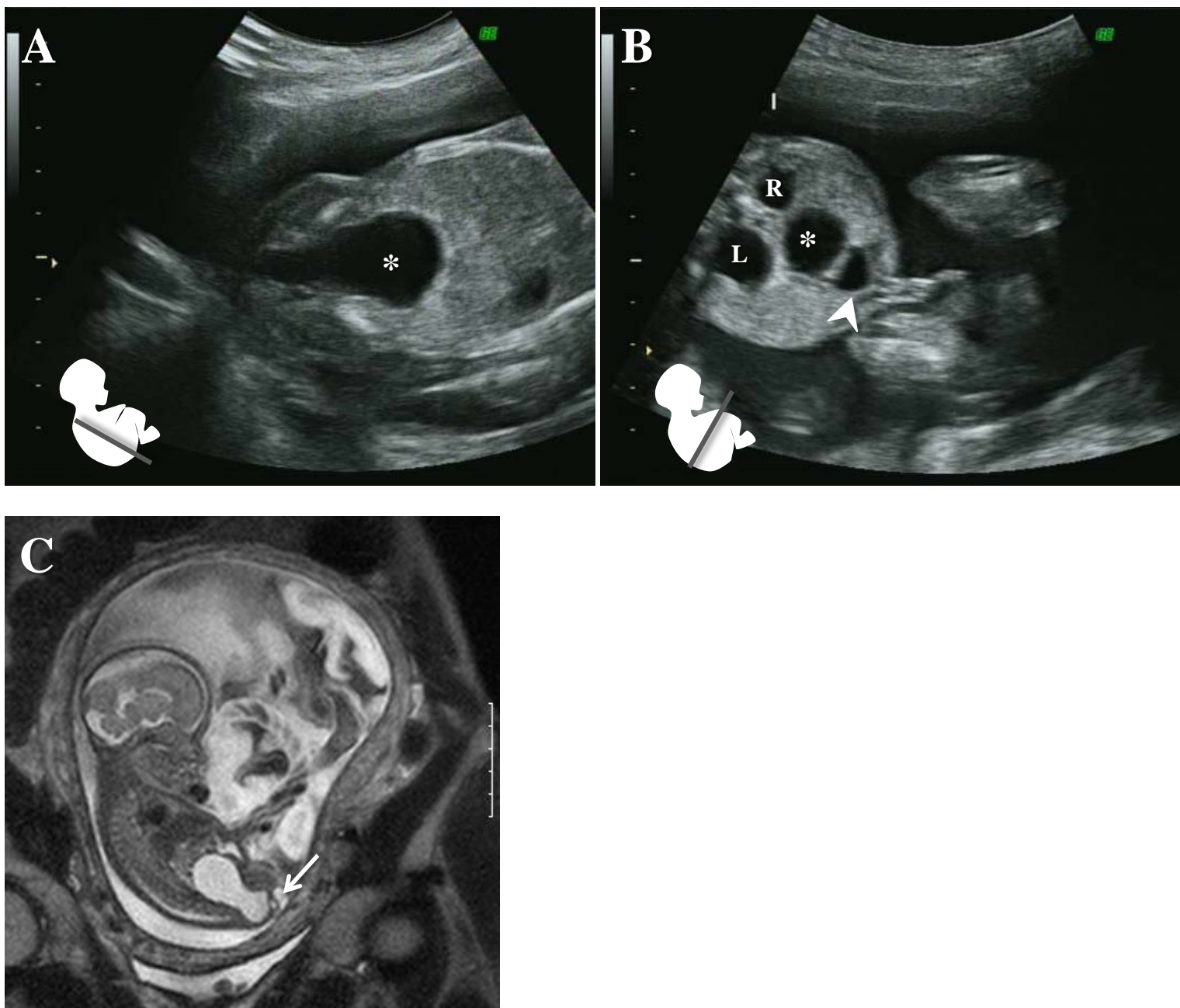
B. Sagittal T2-weighted MRI. Narrow stalk (arrow) connects the pelvic cyst and the spinal canal through the anterior sacral foramen. Multilocular lesion (arrowhead) is located caudally to pelvic cyst.

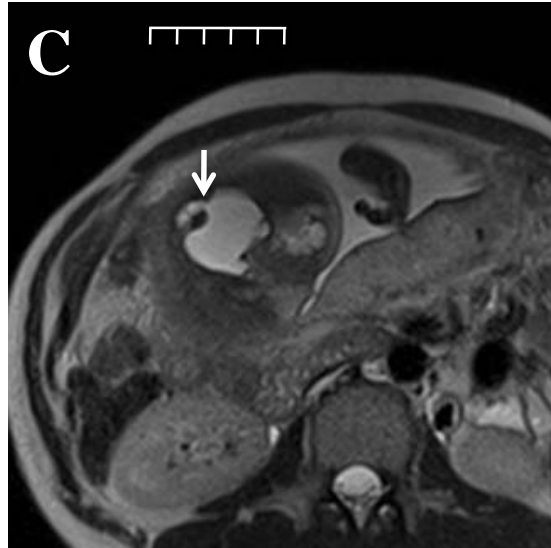
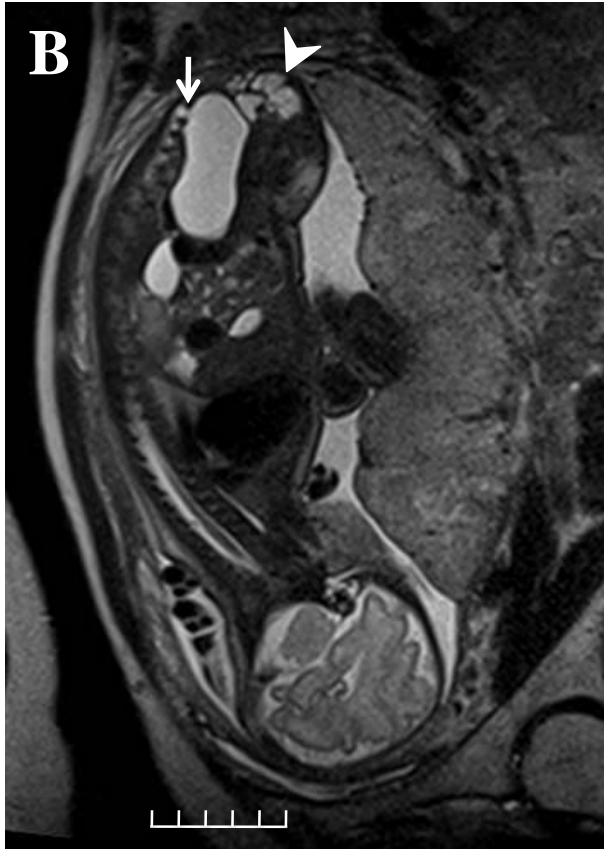
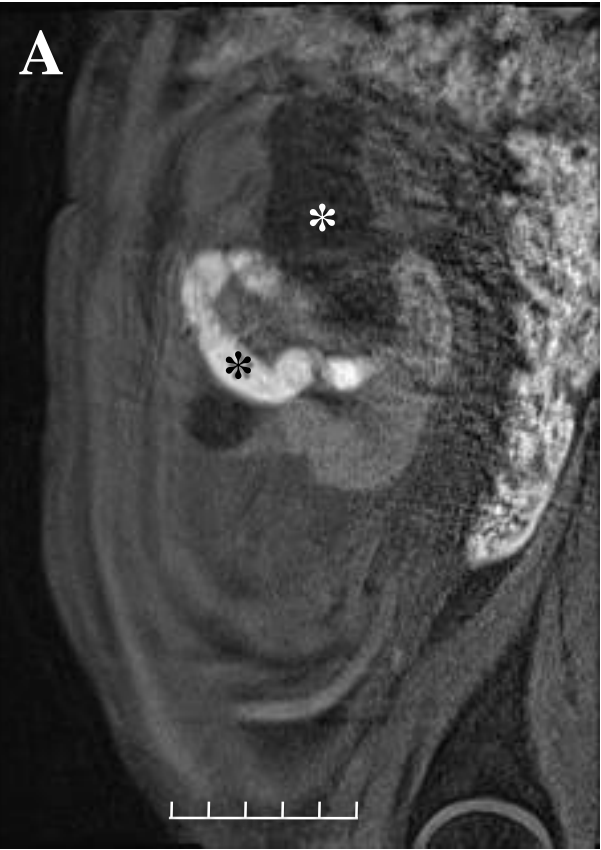
C. Horizontal T2-weighted MRI. Narrow stalk (arrow) connects the pelvic cyst and the spinal canal through the left anterior sacral foramen.

Figure 3. T2-weighted magnetic resonances image at 5 days old (A) and helical computed tomography at 51 days old (B)

A. Huge pelvic cyst is connected with the spinal canal (arrow). Note that caudal multiloculated lesion (arrowhead) is distinct from the pelvic cyst.

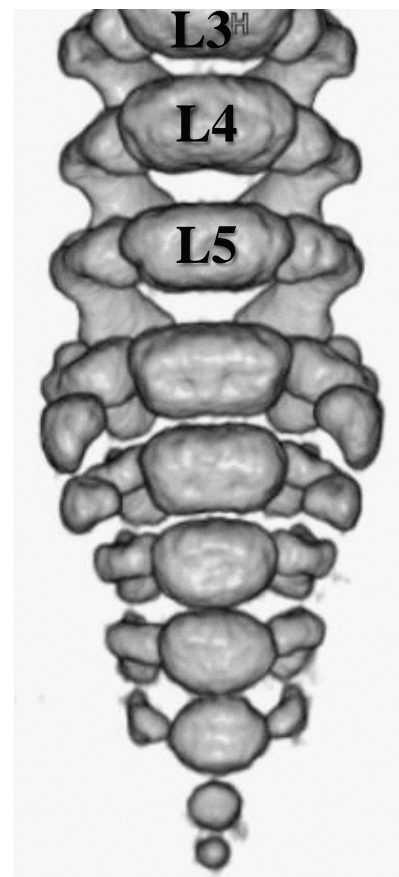
B. Posterior aspect of the vertebral bones below L3 is defective.



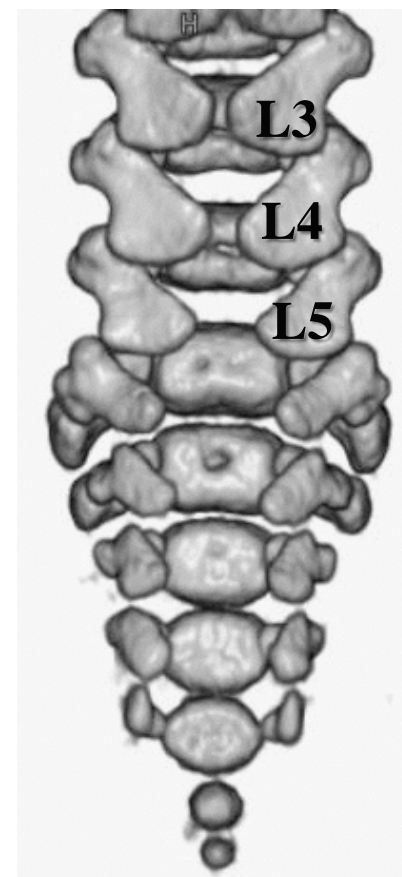




**B**



**anterior aspect**



**posterior aspect**